Hidradenitis suppurativa

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THE REFINED HURLEY QUESTIONNAIRE: AN ACCURATE SELF-ASSESSMENT INSTRUMENT FOR DERIVING THE CORRECT REFINED HURLEY STAGE IN HIDRADENITIS SUPPURATIVA

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INTRODUCTION

Hidradenitis suppurativa (HS) is a chronic, debilitating, inflammatory skin disease that mainly affects body folds (e.g. axillae and groin).[1] Primary lesions include deep-seated inflammatory nodules, abscesses, and sinus tracts.[1] The diagnosis of HS can be made easily due to its clear, distinct clinical presentation.

Deckers *et al.* have reported that patients with HS can score their own disease severity according to the original Hurley classification.[2] However, the purpose of the Hurley classification was to assess HS in a single affected body region in order to guide surgical intervention.[3] It was not intended to classify HS disease activity and severity in the whole patient and to assist in extensive treatment plans including, for example, anti-inflammatory options.[3,4]

Therefore, a modification of the Hurley classification was proposed by a Dutch HS expert group in 2017: the "refined Hurley classification".[5] In contrast to the original Hurley classification, the 7-stage refined Hurley classification assesses not only the presence of sinus tracts, but also inflammatory symptoms and the extensiveness of the disease.[3,5] Refined Hurley stages I and II are subdivided into A, B and C, corresponding to mild, moderate and severe disease. Stage III is not subdivided and corresponds to severe HS disease. This was recently confirmed by a construct validation study.[6] Furthermore, a comprehensive treatment ladder is added to the flow chart.[5]

The aim of this study was to develop and investigate the reliability of a patient self-assessment questionnaire corresponding to the items addressed in the refined Hurley classification, in order to derive the refined Hurley stage.

METHODS

Subjects and study design

Consecutive patients with HS were recruited at the dermatology outpatient clinic of the University Medical Centre Groningen (UMCG), a tertiary referral centre for HS. Subjects were eligible if they were diagnosed with HS by a dermatologist, were older than 18 years, and were capable of completing the questionnaire in Dutch.

A patient symptom self-assessment questionnaire was developed by HS experts based on the refined Hurley classification flow chart. Following a pilot study performed with 16 patients, several modifications were applied to the concept questionnaire. Patients with HS were requested to complete the definitive questionnaire (the refined Hurley classification questionnaire for patients with HS (Appendix)) before their regular consultation at the dermatologist. The refined Hurley stage was derived following the

flowchart by an investigator (RV).[5] The dermatologists were requested to report a detailed dermatological examination and report the refined Hurley classification, as they also do in daily clinical practice. The percent agreement of data entry between 2 investigators (RV and AR), based on a sample of 10 out of 75 (13.3%) randomly chosen subjects, was 97.2%. No formal sample size calculation can be performed for this type of study. Based on literature on methodology and similar studies in the same field, the aim was to include 75 subjects in the final cohort testing.[2,7] For this type of study, medical ethics committee approval is not required under Dutch law.

Statistical analysis

Descriptive statistics were used to describe the study population. The inter-rater agreement and reliability between the HS patient's derived refined Hurley stages and physician's reported refined Hurley stages were calculated. Next, the inter-rater agreement and reliability of the presence of sinus tracts and HS disease severity, defined by the refined Hurley classification, was calculated. For the inter-rater agreement, percentages of agreement between physicians and patients were calculated manually. Because the refined Hurley classification is a nominal scale, a Krippendorff's alpha (α) is suitable to determine the inter-rater reliability.[8] Statistical analysis was performed using IBM SPSS Statistics 23.0 for Windows (SPSS, Chicago, USA). *p*-values ≤ 0.05 were considered statistically significant.

RESULTS

A total of 75 subjects participated in this study. An overview of the patients' characteristics is shown in Table I. Approximately one-third were patients visiting the dermatology outpatients clinic (UMCG) for the first time.

Inter-rater agreement and reliability

The derived refined Hurley stages and disease severity (based on the refined Hurley classification) from the patient's answers to the questionnaire vs. the physician's dermatological examination report are shown in Table SI and Table SII. The inter-rater agreement between patient's derived and physician's reported refined Hurley stages was 78.7% (59/75). The inter-rater reliability resulted in an α of 0.737 (95% confidence interval [CI] 0.622–0.852) (Table SIII). Similar results were found for inter-rater agreement and reliability regarding HS disease severity (82.7%, α=0.733 (95% CI 0.589–0.856) (Table SIII). Concerning the assessment of sinus tracts, inter-rater agreement was 89.2% and reliability of α=0.785 (95% CI 0.650–0.919).

Table 1. Patients' characteristics (n = 75)

Characteristics		
Age, years, *mean ± standard deviation*	40.5	± 12.7
Female sex, *%*	72.0	
Body Mass Index, kg/m^2, *mean ± standard deviation*	29.4	± 6.0
Smoking status, *n* (%)		
Non-smoker	13	(17.3)
Ex-smoker	19	(25.3)
Current smoker	43	(57.3)
New (first visit) or control patient, *n* (%)		
New	26	(34.7)
Control	49	(65.3)
Refined Hurley classification stage, according to physician, *n* (%)		
Refined Hurley IA	22	(29)
Refined Hurley IB	5	(7)
Refined Hurley IC	8	(11)
Refined Hurley IIA	8	(11)
Refined Hurley IIB	11	(15)
Refined Hurley IIC	16	(21)
Refined Hurley III	5	(7)

DISCUSSION

In this study, we developed a patient symptom self-assessment questionnaire based on the refined Hurley classification algorithm for HS. We investigated whether the derived refined Hurley stages from the patient questionnaire correspond to the physician's dermatological examination and given refined Hurley stage. It was found that a *substantial* inter-rater agreement and reliability, indicating that, in most cases, the same refined Hurley stage could be extracted from the patients' answers to our questionnaire as assigned by the physician.

Notably, in contrast to the flow chart of the refined Hurley classification, we found in the current study that it is important to first ask patients with HS about the presence of abscesses/inflammatory nodules, prior to the presence of sinus tracts. This might be due to the chronological order in which HS mostly develops: the first signs of HS are usually recurrent inflammatory nodules and/or abscesses, and in a later stage sinus tracts might develop. Furthermore, the reliability of the questionnaire is enhanced by educating the patient about the main HS lesions, by providing a concise description with prototypical pictures of these lesions.

One of the main items in the original as well as in the refined Hurley classification that has to be determined is the presence of sinus tracts. We have shown that the inter-rater agreement and reliability regarding the presence of sinus tracts is especially high. However, as stated previously, the original Hurley classification lacks valuable information to assess symptoms and severity in an entire individual.[4] Recently, we have shown that the

sub-stages of the refined Hurley classification correlated significantly with patient-reported quality of life and physician-assessed disease severity.[6] In the current study we showed that patients and physicians also agree on the level of disease severity.

Furthermore, compared with the study by Deckers *et al.* and another study regarding self-assessment of disease severity of other skin diseases (acne, psoriasis, and atopic eczema), our results are the highest.[2,9]

A limitation of the current study is that it was conducted in a single university hospital with HS expertise. This might have biased the results. Patients with HS seen at our department might have a longer duration of disease and are usually extensively informed about their disease. This could indicate that these patients are more familiar with the symptoms of HS than are patients treated in primary and secondary healthcare centres. However, besides inclusion of patients coming for follow-up consultation, new referrals were also included.

In conclusion, the symptom self-assessment questionnaire described here is an accurate instrument for deriving the correct refined Hurley stage within patients with HS and might be useful for daily clinical practice, as well as for future epidemiological and clinical studies in HS. We recommend investigating the usefulness of this questionnaire further in other/multiple treatment centers, including sub-analyses, such as the results of new vs. follow-up patients, presence of inflammatory nodules/abscesses, and involved anatomical region.

Acknowledgements

The authors are grateful for the participation of all the patients with HS in this study.

SUPPLEMENT

Table SI. Refined Hurley classification derived from patient's answers to the questionnaire vs. physician's report

		By physician						
		IA	**IB**	**IC**	**IIA**	**IIB**	**IIC**	**III**
By patient	**IA**	21	0	1	1	0	2	0
	IB	0	4	0	0	0	0	0
	IC	0	0	5	0	0	0	0
	IIA	0	0	0	5	1	2	0
	IIB	1	0	0	1	8	2	0
	IIC	0	1	2	1	2	10	0
	III	0	0	0	0	0	0	5

Table SII. Disease severity based on refined Hurley classification derived from patient's answers to the questionnaire vs. physician's report

		By physician		
		Mild (IA+IIA)	**Moderate (IB+IIB)**	**Severe (IC+IIC+III)**
By patient	**Mild (IA+IIA)**	27	2	1
	Moderate (IB+IIB)	1	12	3
	Severe (IC+IIC+III)	4	2	23

Table SIII. Results definitive questionnaire (n=75)

	Inter-rater agreement, %	**Inter-rater reliability, α**	**95% confidence interval**
Derived refined Hurley stage	78.7	0.737	0.622-0.852
Presence of sinus tracts	89.2	0.785	0.650-0.919
Derived disease severity based on refined Hurley classification	82.7	0.733	0.589-0.856

APPENDIX

Definitive questionnaire: "The refined Hurley classification questionnaire for patients with HS"

Vragenlijst Dermatologie

Invuldatum (dd/mm/jjjj)	.. .. / .. .. / .. ..
Uw achternaam	……
Uw geboortedatum (dd/mm/jjjj)	.. .. / .. .. / .. ..
Uw geslacht	☐ Man ☐ Vrouw
Uw lengte	……..meter
Uw gewicht	……..kg
Rookt u?	☐ Ja, circa …… sigaretten per dag sinds het jaartal ……..
	☐ Gestopt, circa ……. jaar ……. sigaretten per dag gerookt.
	☐ Nee, ik heb nooit gerookt.

Deze vragenlijst gaat over Uw huidklachten van hidradenitis suppurativa op dit moment.

Het invullen van de vragenlijst duurt enkele minuten.

Alvast heel hartelijk dank voor Uw medewerking!

Hidradenitis suppurativa

Korte beschrijving van mogelijke klachten die gepaard kunnen gaan met hidradenitis suppurativa

Hidradenitis suppurativa, soms ook HS, acne inversa of acne ectopica genoemd, is een chronische huidziekte. Kenmerkend voor hidradenitis suppurativa zijn terugkerende, pijnlijke ontstekingen in lichaamsplooien, zoals de liezen en/of de oksels. Ook kan men ontstekingen hebben onder de borsten, op de billen, in de schaamstreek/geslachtsdeel en gezicht/oren/behaarde hoofdhuid.
De type ontstekingsverschijnselen kunnen onderverdeeld worden in 2 groepen:

1) De **losse bulten/ontstekingen** liggen meestal diep onder de huid maar kunnen ook lijken op puisten. Ook kunnen abcessen (holtes met pus) ontstaan.

2) Op langere termijn kunnen er **onderhuidse tunnels** ontstaan (ook wel sinussen genoemd). Deze tunnels kunnen in lengte variëren, van ongeveer 1 cm tot meer dan 10 cm.

VRAAG 1 – Losse ontstekingen

Deze vraag gaat over losse ontstekingen. Dit zijn vaak gevoelige/pijnlijke bulten, zoals abcessen, grote puisten en/of rode bulten.

Let op!: hier wordt **niet** gevraagd naar onderhuidse tunnels (sinussen); dit komt bij vraag 2 aan bod.

In afbeelding 1 hieronder ziet u enkele voorbeelden van hoe losse ontstekingen eruit kunnen zien bij hidradenitis suppurativa.

Afbeelding 1. Voorbeelden van **losse ontstekingen** in de oksel bij hidradenitis suppurativa.

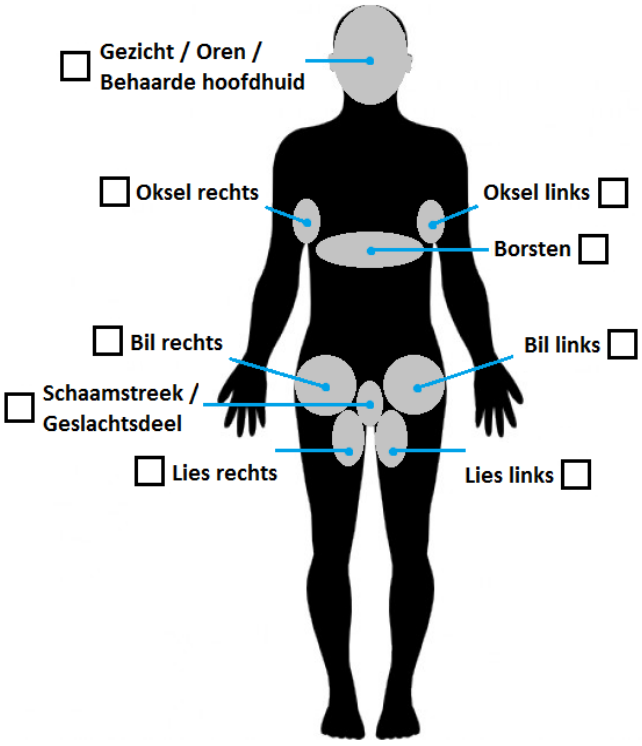
1a. Oksel met een rode bult/abces.
1b. Oksel met een rode bult/abces en twee paarse plekken waarbij de ontsteking al voorbij is.
1c. Oksel met meerdere bulten/abcessen.

1a. Heeft U **op dit moment** ergens op Uw lichaam, zoals in de oksels, liezen, onder de borsten, op de billen, in de schaamstreek en/of gezicht last van **losse ontstekingen**?

- □ Nee, ik heb op dit moment geen losse ontstekingen → *Ga dan door met vraag 2.*
- □ Ja, ik heb op dit moment losse ontstekingen

1b. In welke **lichaamslocaties** heeft U **<u>op dit moment</u> losse ontstekingen**?

U kunt hieronder in de figuur aankruisen in welke lichaamslocaties de losse ontstekingen voorkomen:



1c. Hoeveel losse ontstekingen telt U **<u>op dit moment</u>** in totaal?

- □ Minder dan 5
- □ 5 of meer

1d. Ontstaan de losse ontstekingen voornamelijk op precies dezelfde plaats(en) binnen dezelfde locatie(s), of elke keer op nieuwe/wisselende plaatsen/locaties?

- □ Voornamelijk op dezelfde plaatsen
- □ Voornamelijk op nieuwe/wisselende plaatsen

VRAAG 2 – Onderhuidse tunnels

Deze vraag gaat over onderhuidse tunnels (ook wel sinussen genoemd). Onderhuidse tunnels zijn meestal duidelijk met het blote oog te zien aan het oppervlakte van de huid. De tunnels kunnen ontstoken zijn, dan lekt er vaak pus. Onderhuidse tunnels zullen nooit uit zichzelf verdwijnen. Daarom wordt vaak voorgesteld om deze tunnels chirurgisch weg te snijden.

In afbeelding 2 hieronder ziet u enkele voorbeelden van hoe onderhuidse tunnels eruit kunnen zien bij hidradenitis suppurativa.

Afbeelding 2. Voorbeelden van tunnels bij hidradenitis suppurativa in een oksel. Er is sprake van onderhuidse tunnelvorming, dit is te zien aan de plooien die zich in de huid vormen.

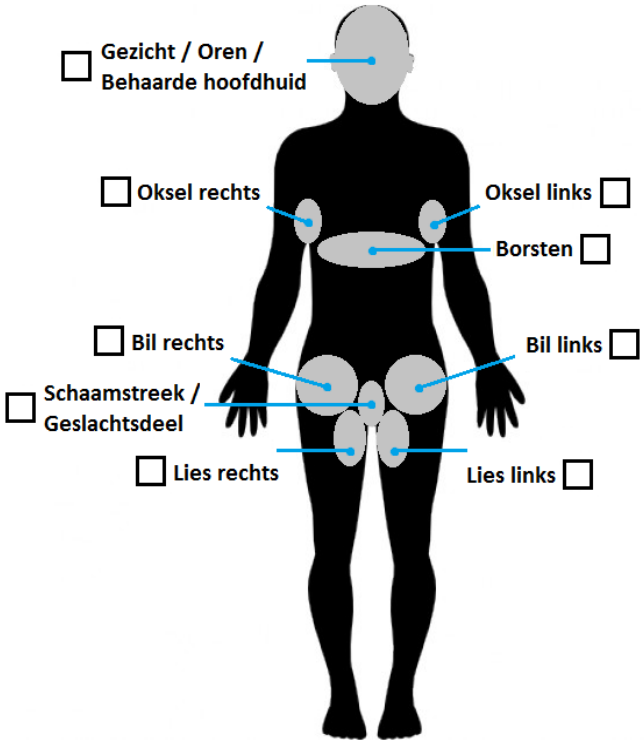
2a. Oksel met beginnende tunnelvorming, zonder ontstekingsverschijnselen.
2b. Oksel met iets uitgebreidere en duidelijkere tunnelvorming, zonder ontstekingsverschijnselen.
2c. Oksel met meerdere tunnels en ontstekingsverschijnselen.

2a. Heeft U **<u>op dit moment</u>** ergens op Uw lichaam, zoals in de oksels, liezen, onder de borsten, op de billen, in de schaamstreek en/of gezicht last van **onderhuidse tunnels**?

- □ Nee, ik heb op dit moment geen onderhuidse tunnels → *Einde vragenlijst*
- □ Ja, ik heb op dit moment onderhuidse tunnels

2b. In welke lichaamslocaties komen de **onderhuidse tunnels** bij U **<u>op dit moment</u>** voor?

U kunt hieronder in de figuur aankruisen in welke lichaamslocaties onderhuidse tunnels voorkomen:



2c. Is het aangedane gebied in één lichaamslocatie waar onderhuidse tunnels voorkomen kleiner óf groter dan het oppervlakte van Uw hand (handpalm plus vingers, zie de afbeelding hiernaast)?

- □ Kleiner
- □ Groter

2d. Zijn één of meerdere van deze onderhuidse tunnels **<u>op dit moment</u>** **gevoelig/pijnlijk én rood**?

- □ Ja
- □ Nee

2e. Lekt er bij één of meerdere van deze onderhuidse tunnels **<u>op dit moment</u>** **pus**?

- □ Ja
- □ Nee

Hartelijk dank voor het invullen van de vragenlijst!

References

1. Jemec GB. Hidradenitis suppurativa. *N Engl J Med.* 2012;366: 158–164.
2. Deckers I, Mihajlovic D, Prens E, Boer J. Hidradenitis suppurativa: a pilot study to determine the capability of patients to self-assess their Hurley stage. *Br J Dermatol.* 2015; 172: 1418–1419.
3. Hurley HJ. Axillary hyperhidrosis, apocrine bromhidrosis, hidradenitis suppurativa, and familial benign pemphigus: surgical approach. *Dermatologic surgery.* New York: Marcel Dekker. 1989:729–739.
4. Revuz JE, Jemec GB. Diagnosing hidradenitis suppurativa. *Dermatol Clin*. 2016; 34:1–5.
5. Horváth B, Janse IC, Blok JL, Driessen RJ, Boer J, Mekkes JR, et al. Hurley staging refined: a proposal by the dutch hidradenitis suppurativa expert group. *Acta Derm Venereol.* 2017; 97:412–413.
6. Rondags A, van Straalen KR, van Hasselt JR, et al. Correlation of the refined Hurley classification for hidradenitis suppurativa with patient reported quality of life and objective disease severity assessment. *Br J Dermatol*. 2019;180(5):1214-1220.
7. Zapf A, Castell S, Morawietz L, Karch A. Measuring inter-rater reliability for nominal data – which coefficients and confidence intervals are appropriate? *BMC Med Res Methodol*. 2016;16:93.
8. Krippendorff K. Agreement and information in the reliability of coding. *Communic Methods Meas.* 2011;5:93–112.
9. Magin PJ, Pond CD, Smith WT, Watson AB, Goode SM. Correlation and agreement of self-assessed and objective skin disease severity in a cross-sectional study of patients with acne, psoriasis, and atopic eczema. *Int J Dermatol.* 2011;50:1486–1490.